Is superior canal dehiscence congenital or acquired? A case report and review of the literature

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Abstract: This article presents a detailed case report of a patient who was diagnosed with superior canal dehiscence at 37 years of age, but who had a suspicious history for that syndrome from at least 10 years of age. The authors hypothesize several reasons for this late diagnosis, with the goal of helping pediatricians, otolaryngologists, and neurologists consider this syndrome in their differential diagnosis of children, adolescents, or adults experiencing dizziness.

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Is superior canal dehiscence congenital or acquired? A case report and review of the literature.

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Abstract

Superior canal dehiscence syndrome (SCDS) is typically diagnosed in adulthood and not in children or adolescents, but it is believed that it is caused by a developmental or congenital anomaly. This theory would predict more diagnoses earlier in live, but until now the facts do not show that. We report a patient, in whom the diagnosis was made at age 37 but who had a suspicious history for that syndrome from at least age 10. We hypothesize some reasons for this late diagnosis and hope this case may have the effect, to make even pediatricians, ENT-doctors and neurologists seeing children think at this syndrome and to help some of their patients, to have a better live.

Introduction

Typical symptoms for SCDS, first described by Minor et al. (1), are hyperacusis for bone conducted sounds (often autophony) and vertigo, induced by loud sounds (Tullio phenomenon) or by increased pressure in the middle ear (Hennebert sign) or in the head (e.g. during sports). Onset of SCDS symptoms and signs are typically in adulthood (2). Nevertheless, a developmental or congenital anomaly is suspected due to thinning of the thickness of bone overlying the intact superior semicircular canal (sSCC) as shown by CT and temporal bone dissection studies (3, 4). This thin bone could presumably be disrupted by trauma or erosion due to pressure of the overlying temporal lobe (3), but a long term study about development of SCDS on the intact side by follow up with CT or VEMPs has not been reported. A genetic defect could not yet been found (Roknic et al, 2008). Besides trauma other causes could theoretically cause SCDS or other perilymphatic fistulas like cholesteatoma (7), vestibular schwannoma (3), meningioma (8) or even glioblastoma (9), but these are rarely found with SCDS, so other causes seem to be more likely for the majority of cases. Only two reports of a 7 year old girl with SCDS on the right (5) and of a 4 year old child with a partial dehiscence of the right posterior canal (6) have been published. It appears presently that SCDS in children is either rare, underdiagnosed or not published. We believe that a developmental deficit is the most likely cause for SCDS, but why the diagnosis is usually first made in adulthood is still a question.

Case report

We present a patient in whom SCDS was diagnosed at age 37 with typical symptoms. He complained about periods of dizziness, usually provoked by loud sounds. Spells had increased since age 34, and also his sensitivity to noise and autophonia. He even heard his own eye movements in his right ear. Dizziness was increased by exertion in sport activities so much that he could not even go jogging after the age of 35. He never experienced rotatory vertigo.

Otoscopy was normal, Weber lateralized to the right, Rinne was bilaterally positive. He felt a slight shift of the visual scene and a little dizziness during Valsalva maneuver. Hennebert's signs was positive with

small torsional movements of the eyes towards left and back while pressure was applied or released, respectively. This simple investigation was sufficient to suspect SCDS.

Audiometry showed normal hearing with an air bone gap of 15db at 500 Hz and cervical vestibular evoked myogenic potentials (VEMPs) confirmed a reduced threshold of about 46 db nHL on the right and a normal threshold (82 db) on the left (Fig.1).

Fig. 1 about here

As the final proof a CT of the temporal bones showed the expected dehiscence of the right superior canal.

In a more detailed history we found that even as a child, beginning at about 10 years of age, he felt periodically vague dizziness, especially in relatively noisy public places. The complaints also increased with exertion in sporting activities, especially biking uphill. He felt best when lying in bed in a quiet surround. He avoided common activities together with friends, because these were too loud for him. He often told his parents, that something must be wrong with him and was evaluated by several doctors. These presumed multiple diagnoses like vitamin deficiencies and also psychogenic causes (phobic vertigo), but no therapy helped. The patient learned to live with these symptoms.

When we saw the patient, there were no indications of panic disorders, depression, aggravation or any other psychogenic disturbances. After he knew the diagnosis and the operative treatment option, he waited 3 more years with increasing symptoms before he decided upon surgery. The dehiscent canal was plugged via middle cranial fossa approach without complications. Within 48 hours after surgery the patient noted profound changes, even commenting on what seemed like a “new life.”

Discussion

The adulthood presentation of this case appeared quite typical for SCDS. Nevertheless, the patient’s recollections suggest that the right superior canal was dehiscent from childhood because the major symptom (dizziness) already appeared. Delays in making this diagnosis are common (10), in part because SCDS is still a diagnosis not familiar to clinicians outside of those subspecialties dealing commonly with vertigo. Another reason for the delay may be patients reluctance to complain openly of seemingly bizarre symptoms such as hearing one’s own eyes move. This case, however, points out that the diagnosis may be delayed from childhood to adulthood.

As a child, our patient experienced vague symptoms that he found difficult to communicate to his parents and that seemed to be absent in his peers. Unable to understand these symptoms or relate them to others, the patient simply adjusted his activities to avoid the noisy environments or straining that provoked them. Over time, much of this avoidance became unconscious habit incorporated into his otherwise successful lifestyle.

However, for some unknown reason his symptoms worsened as an adult and led him to seek an explanation and treatment. Maybe the dehiscence began to transmit more pressure between the inner

ear and intracranial space as the patient aged as might occur by changing elasticity of the dura, or because the pressure gradients between the inner ear and intracranial space changed. A loss of central compensatory mechanisms having minimized symptoms seems unlikely, because of immediate recovery from all symptoms shortly after the operation. This strongly suggests some change in the physiology of the dehiscence to explain the worsening of symptoms in adulthood. The nature of this change is yet to be determined.

This case raises the possibility that SCDS symptoms may be present in childhood and may go unrecognized. Increased awareness of this diagnosis may uncover more children with SCDS. A high degree of suspicion for SCDS should be maintained by those who evaluate young dizzy patients. Symptoms such as autophony may seem so out of the ordinary to children that they may not spontaneously offer these complaints to their parents or clinicians. A careful history is essential to uncovering the diagnosis of SCDS, and the clinician may have to inquire about apparent agoraphobia to find if it is really a manifestation of situational dizziness provoked by noisy, crowded places. Furthermore, the physical examination (see above) and appropriate interpretation of audiometric findings provides further clues. Suggestive findings from the history or physical exam should prompt further testing, including VEMP testing and high-resolution CT scanning of the temporal bones with appropriate reconstructions of the superior canal (11).

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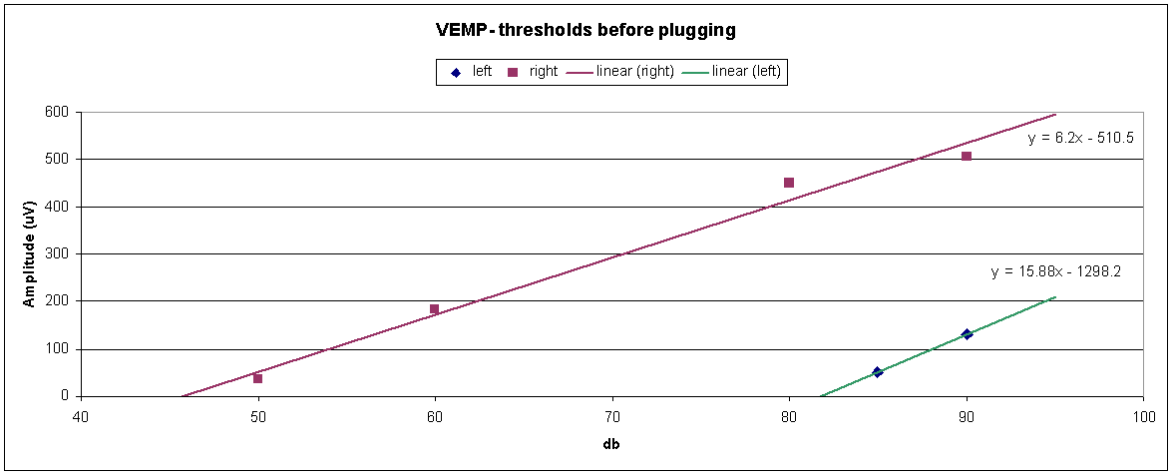
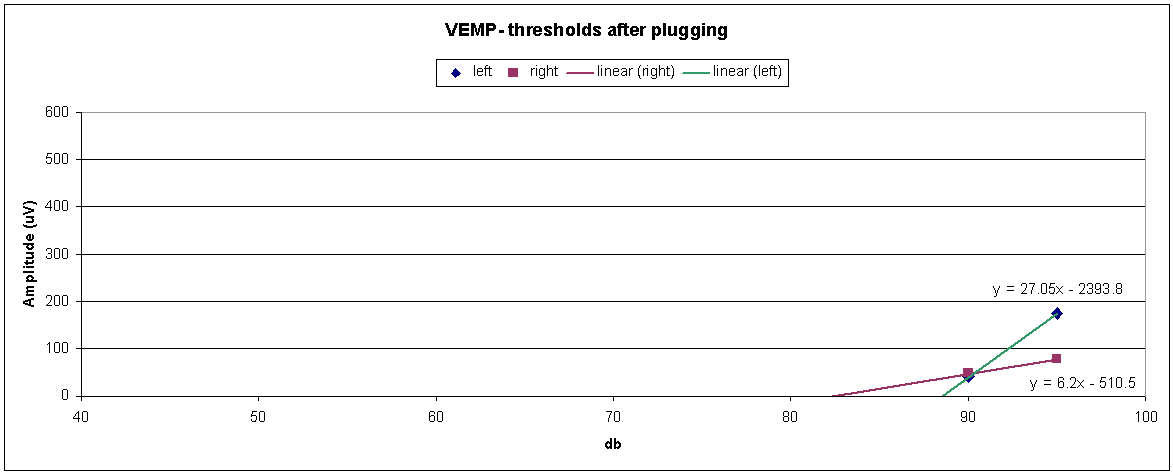
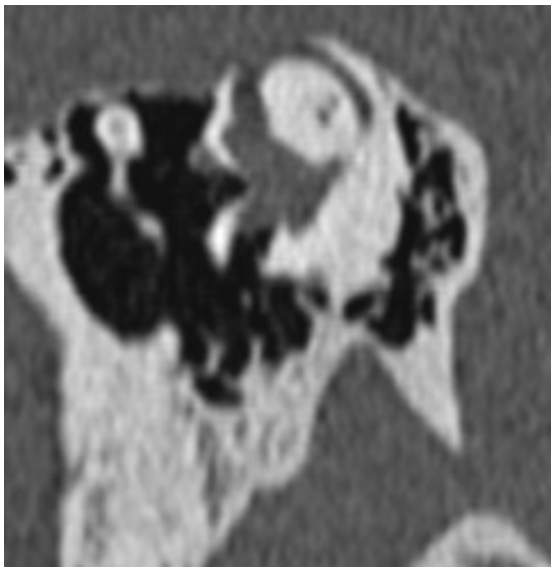
Legends

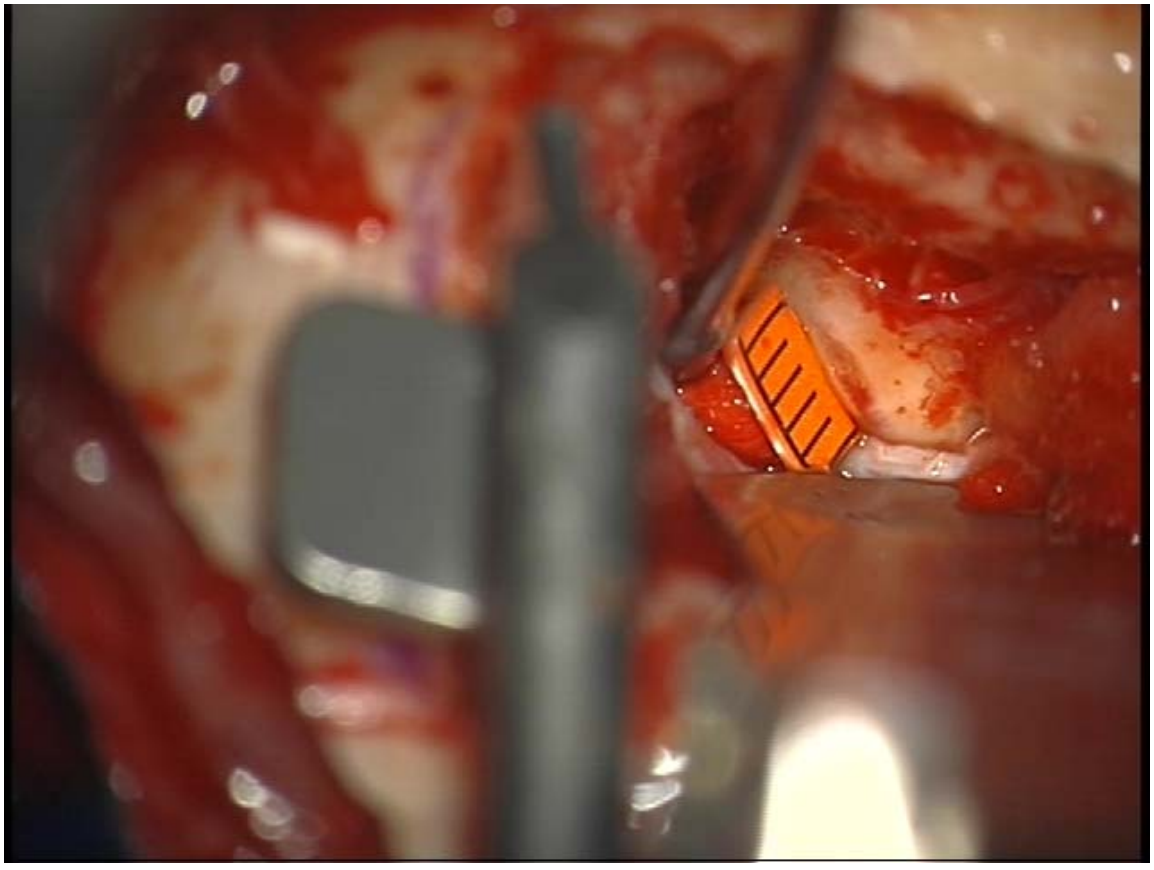
Fig 1: showing the VEMP thresholds before plugging of the right superior canal. The threshold is determined by a linear regression through the amplitudes of the potentials at different stimulus intensities. It is a very rough way to determine the threshold but since the change in amplitude with stimulus intensity is usually rather linear, we often use only two stimulus intensities, if the threshold has normal values. As can be seen, even with 4 intensities, used in A for the threshold of the right sacculus, the amplitudes lie close to the linear regression and the threshold is at about 46 db which is massively below normal, which is > 70 db in our lab.







CT_SCDS right



SCDS intraop.